Published in final edited form as:
Am J Hosp Palliat Care. 2009 ; 26(5): 405–412. doi:10.1177/1049909109346165.

Pain in Youths With Neuromuscular Disease

Joyce M. Engel, OT, PhD, **Deborah Kartin, PT, PhD**, **Gregory T. Carter, MD, MS**, **Mark P. Jensen, PhD**, and **Kenneth M. Jaffe, MD**
Department of Rehabilitation Medicine, University of Washington School of Medicine, Seattle, Washington

Abstract

To examine the prevalence and characteristics of pain in children with neuromuscular disease (NMD), 42 youths with NMD underwent a comprehensive evaluation including a detailed intake interview and structured questionnaire that included demographic and functional data. Youths who reported chronic pain were further queried about pain characteristics, locations, and intensity using an 11-point numerical rating scale and a modified Brief Pain Inventory (BPI). The sample consisted of 24 males (57%) and 18 females (43%), ages ranging from 9 to 20 years (M = 14.8, SD = 2.96). Participants included 14 (37%) with Duchenne-muscular dystrophy, 6 (14%) with myotonic dystrophy, 2 (5%) with Becker dystrophy, 2 (5%) with limb-girdle dystrophy, 2 (5%) with congenital muscular dystrophy, 1 (2%) facioscapulohumeral, and 15 (36%) were classified as "other NMD." Twenty-one (50%) were ambulatory; 26 (62%) used power wheelchairs/scooters, 9 (2%) used manual wheelchairs, 3 (.07%) used crutches/canes, and 1 (2%) used a walker. A total of 23 (55%) of the youths reported having chronic pain. Current pain intensity was 1.30(range=0–6), mean pain intensity over the past week was 2.39 (range = 0–7), mean pain duration was 8.75 hours (SD=12.84). Pain in the legs was most commonly reported and 83% reported using pain medications. This study indicates that chronic pain is a significant problem in youths with NMD. These data strongly support making comprehensive pain assessment and management an integral part of the standard of care for youths with NMD.

Keywords

muscular dystrophy; neuromuscular disease; chronic pain; disability; myopathy

Introduction

Historically pain has not been considered to be a major part of the symptom manifestation of neuromuscular disease (NMD). However there are now a growing number of studies indicating that chronic pain is a common symptom for the majority of people with NMD. Studies of adult persons with a variety of NMDs, including facioscapulohumeral (FSHD), myotonic (MMD), and limb girdle LGMD) forms of muscular dystrophy, the spinal muscular atrophies (SMA), Charcot-Marie-Tooth (CMT) disease, and postpolio syndrome (PPS), have shown reported rates of chronic pain ranging from 70%, to 96%.[1–12] In general, the frequency and intensity of pain reported in adults with NMD is significantly greater than levels of pain reported by the general US population, and comparable to pain reported by adults with osteoarthritis and chronic low back pain.[1–12]

Address correspondence to: Joyce M. Engel, Department of Rehabilitation Medicine, University of Washington, Box 356490, Seattle, WA 98195; jmengel@u.washington.edu.

Recently there has been increasing attention and resources directed towards recognizing, understanding, and managing chronic pain in children with NMD.[13] Zebracki and Drotar recently examined the prevalence and characteristics of pain in children with Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD), including exploring any disagreements regarding pain symptoms among children, parents, and physicians.[14] Their data showed that the majority of boys (54%–80%) with DMD/BMD (these disorders affect males only) reported experiencing pain. Parental reports of pain were higher (70%–90%). Pain typically occurred in the lower back, spine, and legs, and was described as "aching." Both children and parents indicated significantly more intense pain than the physician. This study indicates that pain is not only common in boys with DMD/BMD but is also under recognized by treating physicians.

This is consistent with the frequency of pain reported in other disabling childhood disorders, including cerebral palsy (CP), where the experience of pain continues into the adult years as well.[15–19]

The extent and nature of pain in other forms of childhood NMD has not been well studied. The purpose of this study was to explore the nature and scope of chronic pain in a sample of youths with NMD. Secondary aims included gaining specific knowledge about the nature of pain, including defining the (1) pain frequency, (2) pain intensity, (3) pain location, (4) pain quality, (5) pain interference with function and quality of life (QoL), and (6) pain interventions and their effectiveness.

Methods

Participants

All youths who met the study criteria were invited to participate in this study. The study was approved by the Institutional Review Board of Children's Hospital and Regional Medical Center (CHRMC; Seattle, Washington) and the Muscular Dystrophy Association (MDA) prior to data collection. Youths with NMD and their parents were recruited through mailings from the rehabilitation medicine, neurodevelopmental, and spasticity clinics at CHRMC, a summer camp for youths with NMD, in addition to public postings and word of mouth. Inclusion criteria for youths included (1) primary diagnosis of NMD, (2) chronological age between 8 and 20 years, (3) capable of expressive communication using augmentative communication devices as needed, (4) use of English as the primary language, and (5) no more than mild cognitive impairment as measured on a modified Mini-Mental Status Evaluation (MMSE).[20]

Procedures

Youths completed the questionnaires either in person or over the telephone, depending on the family preference, distance from the university, and speech intelligibility on the telephone. Parents or guardians completed their questionnaires simultaneously during in-person youth interviews or through the mail for those youths doing telephone interviews. At the beginning of the interview, the study was explained and informed parent/guardian consent and youth assent were obtained. For those youths participating by telephone, informed parent/guardian consent and youth assent were obtained prior to the interview.

Measures

A structured 1-time interview questionnaire was developed for the study and consisted of 7 sections:

1. Demographic/descriptive data were collected from parent reports and included sex, age, ethnicity, type of NMD, use of mobility devices and orthotics/splints, use of augmentative or alternative communication devices, and income.

2. Youths who reported recurrent bothersome pain of at least 3 months' duration were then queried about pain characteristics and locations. Pain lasting

3. months or longer is considered *chronic* pain.[21,22] Each participant was asked specific questions about pain locations throughout the body. Pain intensity was rated specific to each pain location on an 11-point numerical rating scale (NRS) from 0 to 10 in which 0 = *no pain* and 10 = *pain as bad as could be*. Pain frequency, pain duration, and exacerbating and relieving pain factors were participant-identified. The numerical rating scale[23,24] has been shown to be reliable and valid for youths as young as 5 years of age. 3. Pain interference was assessed through use of a modified Brief Pain Inventory (BPI).[25–27] The BPI is a valid and reliable instrument designed to measure pain interference.[25–27] Degree of interference with daily activities and participation was measured for the primary pain location. The amount of pain interference in the past week was rated using an 11-point NRS where 0 = *does not interfere* and 10 = *interferes completely*. Since many persons with physical disabilities may be nonambulatory, interference with "ability to get around" replaced the original question of interference with "ability to walk." One question was slightly modified for content to make it more age appropriate (ie, "normal work" was changed to "school or play"). To obtain more information on activities and participation, the investigators added items for interference with self-care, social activities, and recreational activities, communication with others, and learning new information or skills.

4. A parent described the child's level of gross motor function using descriptive categories based on a modified Gross Motor Function Classification System(GMFCS).[28] The original GMFCS is a reliable and valid standardized rating system for children with CP to provide an objective classification of their gross motor function. The modification was the addition of the category "walks without restrictions and has no limitations in gross motor skills."

5. Youth responded to the Functional Disability Inventory (FDI) to provide information on disruption of typical physical and social activities, including schoolwork, due to illness, pain, or disability. [29] The FDI is comprised of 15 behaviors related to functioning in child-relevant settings. For example, "In the last few days, would you have had any physical trouble or difficulty doing these activities (eg, being at school all day)?" Participants were provided with a visual scale to rate their responses on a 1-to-5 scale where 1 = *no trouble* and 5 = *impossible*. The FDI has acceptable psychometric properties.[27,29]

6. Youths reported on the average duration of pain over the past 4 weeks in the body location they identified as most painful. Youths were also asked to report on the frequency of pain at this or these pain locations in the past 4 weeks, using a scale of 1 to 4 where 1 = *always*, 2 = *daily*, 3 = *weekly*, and 4 = *monthly*.

7. The parent was asked to identify treatments used for his or her child's pain, using a checklist. The list included physical therapy, transcutaneous electrical nerve stimulation (TENS), therapeutic massage, occupational therapy, counseling or psychotherapy, and medications (eg, narcotics, nonsteroidal anti-inflammatory drugs [NSAIDS], acetaminophen). The parent also had the option of writing in any additional treatments used. The parent also rated the helpfulness of treatment using a 6-point NRS where 0 = *not at all* and 5 = *extremely*, duration of the treatment use, and whether that pain intervention is currently used.

To eliminate potential influence on responding, parents and their children were queried separately about the youths' experience of chronic pain. For the purposes of this study, the frequency of chronic pain reported by the youths themselves was used for data analysis, rather than the frequency of pain reported by the parents, as this was considered to be a more reliable indicator of actual pain experience.[30,31]

Results

Fifty-three parent-and-child dyads were approached. A total of 42 completed parent-and-child (dyad) structured questionnaires met inclusion criteria. Of all the potential participant dyads approached about the study, 12 youths were classified as ineligible; 9 did not pass the modified MMSE and 3 youths were classified by their parents as ineligible (no specific reason given). One youth declined to participate. The mean duration of the interview of the youths with pain was 32 minutes (SD = 13.5 minutes). None of the participants used augmentative or alternative communication devices.

Description of Participants

Parental reports indicated the following. The sample consisted of 24 males (57%) and 18 females (43%) with ages ranging from 9 to 20 years (M = 14.8, SD = 2.96). Most of the participants (n = 36; 86%) were Caucasian, 3 (7%) were African American, (3) 7% were Asian American, 2 (5%) were Hispanic, and 1 (2%) reported "other." The median annual household income was reported as US$40 000 to US$50 000.

Parental reports indicated 14 (37%) had DMD, 6 (14%) had MMD, 2 (5%) BMD, 2 (5%) LGMD, 2 (5%) had congenital muscular dystrophy, 1 (2%) FSHD, and 15 (36%) were classified as "other NMD," which included the CMT syndromes, all forms of SMA, and many forms of mitochondrial and congenital myopathies. Multiple comorbid medical conditions were reported, including asthma (n = 6, 14%), headaches (n = 3, 7%), and recurrent abdominal pain (n = 3, 7%). In addition, 1 child reported insomnia and 1 reported sleep apnea. One child was reported to have Hirschsprung disease. Twelve (29%) youths were reported to have an unspecified learning disability.

Parental reports indicated 25 (60%) of the youths had at least 1 joint contracture. Contractures were reported for all joint locations. The most common contracture locations identified were the ankles (n = 17, 41 %), knees (n = 17, 41%), hips (n = 12, 29%), elbows (n = 11, 26%), spine (n = 8, 19%), wrists (n = 8, 19%), fingers (n = 5, 12%), toes (n = 4, 10%), and shoulders (n = 3, 7%). Many of the youths had orthopedic surgeries, including spinal fusion/stabilization (n = 12, 29%), tendon transfers/lengthening (n = 6, 14%), and contracture release (n = 1, 2%). Three participants (7%) had undergone gastrostomy tube replacement for nutritional support.

In terms of locomotion, 21 (50%) were full-time ambulators without assistive devices. Of the remaining youths, 13 (31%) used power wheelchairs, 4 (10%) used manual wheelchairs, 3 (7%) used crutches/canes, and 1 (2%) used a walker for mobility at home and in the community. We separated data on home and community settings. Splints/orthotics were used by 15 (36%) participants.

Thirty (71%) of the children were reported by their parents to have chronic pain, where as only 23 (55%) of the youths themselves reported having chronic pain. Pain frequency is presented in Table 1. The mean current pain intensity using the 11-point NRS was reported as 1.30 (range = 0–6). The mean pain intensity over the past week was 2.39 (range = 0–7). The reported duration of the pain episodes experienced by the youths ranged from brief

episodes lasting a minute to prolonged episodes of pain lasting as long as 2 days. The mean pain duration was 8.75 hours (SD = 12.84).

Data From NMD Youths Who Reported Chronic Pain

Of the youths who reported chronic pain (n = 23), most experienced pain in multiple body locations. Pain in the legs was most commonly reported. Table 2 displays the frequency and range of pain intensity per body location. Pain interfered with all 12 items on the modified BPI. The range of interference was mild to severe. Table 3 presents data on pain interference for all of the BPI items.

Youths with NMD identified a wide range of exacerbating and relieving pain factors, as detailed in Table 4. As can be seen from Table 4, there was a wide range of factors that exacerbated pain. They included “feeling down,” cold weather or being cold, eating, lifting, prolonged sitting or lying, running, standing, and touch. The most commonly reported factors that helped to relieve pain identified by the youths are presented in Table 5. Lesser common strategies that were identified to help relieve pain included changing body position/posture, swimming, sitting, warmth, changing activity, and wearing socks.

Table 6 details the frequency and helpfulness of pain treatment. A significant number of youths reported using both over-the-counter analgesics (eg, acetaminophen, NSAIDS) and prescription opioid medication.

Discussion

Our study indicates that pain is a commonly experienced symptom in youths with NMD. Over 70% of the parents reported chronic pain in their children with NMD and more than half of the youths self-reported chronic pain. This finding is consistent with previous studies of adults and youths with various disabling conditions, including spinal cord injury, cerebral palsy, amputation, and neurological injuries.[1–9,14,15–18,32–37] Previous studies in adults with NMD show that a significant percentage have substantial problems with pain, severe enough that it negatively affects both functional capacity and QoL.[1–9]

Reported pain intensity ranged from 0 (no pain) to 7 (severe), with the typical pain episode lasting almost 9 hours. Although some pain episodes were of mild intensity, the duration was long. The potential effect of protracted duration of pain, even when the intensity is mild, has implications for youths’ engagement in activities and participation as well as depression, suffering, and overall QoL. Chronic pain has been previously reported as negatively affecting many aspects of youth and adolescent life.[38–41] Our data support this as more than three quarters of the youths reported that pain indeed affected their daily routines, activity, and participation. In addition, a small subsample reported severe pain intensity.

The high percentage of youths with NMD in this sample reporting chronic pain, and in multiple locations, highlights the need for clinicians working with youths with NMD to routinely inquire about the presence of chronic or recurrent pain and the pain locations throughout the entire body. Pain is often not routinely assessed by health care providers. Moreover, studies have shown that physicians frequently underestimate the severity of pain experienced by patients with disabilities, particularly in the context of disability or disruptive behavior, both of which may be present in this population.[42–45] Thus, pain management may not even be considered.

Our data also suggest that multiple methods for pain management may prove beneficial as youths reported using a variety of measures, all affording at least some temporary relief. However, in our study sample, the effects of various strategies to alleviate pain are

inconsistent and appear to be individualized. For example, for some youths exercise exacerbated pain, while for others it alleviated pain. This is similar to the experience of children with CP.[15–18] Given the complexity of all forms of NMD, it is likely that the best pain management strategies would entail an individualized, multimodality approach to have the most chance of success. However, further research is indicated before any specific conclusions can be drawn.

From a clinical perspective, the loss of mobility and physical function is likely playing a major role in the experience and processing of pain in youths with NMD. Coping responses, and other psychosocial factors, have been reported to significantly impact the experience of chronic pain in the adult NMD population.[2] Moreover, measures of psychological functioning and pain interference are associated with multiple psychosocial variables in the adolescent patient population.[46,47] Thus, the adolescent with disability would be at high risk to develop maladaptive pain-coping strategies. Untreated chronic pain could potentially be an underlying cause of antisocial or disruptive behavior that has been reported in this patient population.[13,14,41,45] These behaviors may place enormous stress on interpersonal relationships within the family structure, which is often ill equipped to handle the stresses of caring for a child with disability.[48]

Our data also suggest that sleep and mood disorders are more commonly reported in the youths with NMD who report chronic pain. Nearly 80% of the youths with NMD in our sample reported experiencing some sleep problems. The magnitude of this problem is great and has implications not only on the health and well being of the youths, but on a cascade of effects on the family of the child who experiences chronic sleep problems. Some possible causes for sleep disruption may be related to physical aspects of NMD such as inability or limited ability to adjust one's position while sleeping or airway insufficiency secondary to hypotonia, as reported by the youth with sleep apnea in our sample. Sleep-disordered breathing is very common in the pediatric NMD population, particularly DMD and SMA.[49–58] This is due primarily to weak muscles of inspiration and expiration. [49–58] However, there may also be a component of diminished central drive to breathe in some of the NMDs, most notably MMD and DMD.[49,51] Phrenic nerve dysfunction may also contribute to nocturnal hypoventilation in CMT.[56,58] Additionally, about one quarter of our sample of youths with NMD complained of restless/aching legs. Impaired sleep quality is known to contribute to daytime symptoms of inattention, irritability, poor impulse control, hyperactivity, or poor academic performance in able-bodied adolescents with chronic pain.[59,60] In our study, chronic pain and fatigue could be, at least partially, due to an underlying sleep disorder secondary to nocturnal hypoventilation and this may be an important target for clinical intervention.

Although the qualitative nature of our methodology and lack of a control group limit the extent to which conclusions can be firmly drawn, our data nevertheless clearly indicate that chronic pain is a significant problem for youths with NMDs. The next step would be multicentered studies done with larger numbers of children, each focusing on a single NMD, rather than a diverse mixture of diseases such as was done in this study. Separating out studies into single-disease entities would allow investigators to compare the experiences of children with differing disorders, both myopathic and neuropathic. This would hopefully permit the formulation of better, more specific diagnostic tools and ultimately facilitate the development of effective treatment paradigms. Challenges include dealing with the progressive nature of the vast majority of NMDs, which creates a "moving target" for clinicians trying to provide effective treatment. Thus, future studies would optimally be prospective and longitudinal. This would help best determine the complex interrelationships between chronic pain, physical disability, sleep and mood disorders, and family and interpersonal relationships in persons with NMDs across the lifespan. In the meantime, our

current study, as well as other studies cited herein, provide ample evidence that youths and adults with NMDs do experience significant problems with pain that negatively affect almost every aspect of their lives. Clinicians caring for this patient population need to begin more effectively addressing these issues, perhaps first and foremost by simply inquiring about the nature and extent of pain in the patient with NMD, the nature and extent of their pain. Given the findings in our study, optimal treatment would likely be multimodal, involving not only pharmaceutical agents but also physical rehabilitation and psychosocial interventions.

Acknowledgments

National Institute of Child Health and Human Development Grant number P01 HD33988; and National Institute on Disability and Rehabilitation Research Training Center Grant number HB133B980008.

References

1. Abresch RT, Carter GT, Jensen MP, Kilmer DD. Assessment of pain and health-related quality of life in slowly progressive neuromuscular disease. Am J Hosp Palliat Care 2002;19(1):39–48. [PubMed: 12173612]
2. Miro J, Raichle KA, Carter GT, et al. Impact of biopsychosocial factors on chronic pain in persons with myotonic and facioscapulohumeral muscular dystrophy. Am J Hosp Palliat Med. 2009 Epub ahead of print.
3. Jensen MP, Hoffman AJ, Stoelb BL, Abresch RT, Carter GT, McDonald CM. Chronic pain in persons with myotonic and facioscapulohumeral muscular dystrophy. Arch Phys Med Rehabil 2008;89(2):320–328. [PubMed: 18226657]
4. Jensen MP, Abresch RT, Carter GT, McDonald CM. Chronic pain in persons with neuromuscular disorders. Arch Phys Med Rehabil 2005;86(6):1155–1163. [PubMed: 15954054]
5. Carter GT, Jensen MP, Stoelb BL, Hoffman AJ, Abresch RT, McDonald CM. Chronic pain in persons with myotonic muscular dystrophy, type 1. Arch Phys Med Rehabil 2008;89(12):2382. [PubMed: 19061752]
6. Carter GT, Jensen MP, Galer BS, et al. Neuropathic Pain in Charcot Marie Tooth disease. Arch Phys Med Rehabil 1998;79(12):1560–1564. [PubMed: 9862301]
7. Stoelb BL, Carter GT, Abresch RT, Purekal S, McDonald CM, Jensen MP. Pain in persons with postpolio syndrome: frequency, intensity, and impact. Arch Phys Med Rehabil 2008;89(10):1933–1940. [PubMed: 18929021]
8. Jensen MP, Abresch RT, Carter GT. The reliability and validity of a self-reported version of the functional independence measure in persons with neuromuscular disease and chronic pain. Arch Phys Med Rehabil 2005;86(1):116–122. [PubMed: 15641001]
9. Hoffman AJ, Jensen MP, Abresch RT, Carter GT. Chronic pain in persons with neuromuscular disorders. Phys Med Rehabil Clin N Am 2005;16(4):1099–1112. xii. [PubMed: 16214063]
10. Carter GT, Han JJ, Abresch RT, Jensen MP. The importance of assessing quality of life in patients with neuromuscular disease. Am J Hosp Palliat Med 2007;23(6):493–497.
11. Abresch RT, Jensen MP, Carter GT. Health quality of life in peripheral neuropathy. Phys Med Rehabil Clin N Am 2001;12(2):461–472. [PubMed: 11345018]
12. Tiffreau V, Viet G, Thevenon A. Pain and neuromuscular disease: the results of a survey. Am J Phys Med Rehabil 2006;85(9):756–766. [PubMed: 16924188]
13. Engel JM, Kartin D, Jaffe KM. Exploring chronic pain in youths with Duchenne Muscular Dystrophy: a model for pediatric neuromuscular disease. Phys Med Rehabil Clin N Am 2005;16(4):1113–1124. [PubMed: 16214064]
14. Zebracki K, Drotar D. Pain and activity limitations in children with Duchenne or Becker muscular dystrophy. Dev Med Child Neurol 2008;50(7):546–552. [PubMed: 18611207]
15. Engel JM, Jensen MP, Schwartz L. Coping with chronic pain associated with cerebral palsy. Occup Ther Int 2006;13(4):224–233. [PubMed: 17623374]

16. Jensen MP, Engel JM, Schwartz L. Coping with cerebral palsy pain: a preliminary longitudinal study. Pain Med 2006;7(1):30–37. [PubMed: 16533194]
17. Engel JM, Petrina TJ, Dudgeon BJ, McKearnan KA. Cerebral palsy and chronic pain: a descriptive study of children and adolescents. Phys Occup Ther Pediatr 2005;25(4):73–84. [PubMed: 16418117]
18. Jensen MP, Engel JM, Hoffman AJ, Schwartz L. Natural history of chronic pain and pain treatment in adults with cerebral palsy. Am J Phys Med Rehabil 2004;83(6):439–445. [PubMed: 15166688]
19. Jensen MP, Engel JM, McKearnan KA, Hoffman AJ. Validity of pain intensity assessment in persons with cerebral palsy: a comparison of six scales. J Pain 2003;4(2):56–63. [PubMed: 14622716]
20. Folstein M, Folstein SE, McHugh PR. “Mini-Mental State”. A practical method for grading the cognitive state of patients for the clinician. J Psychiatr Res 1975;12(3):189–198. [PubMed: 1202204]
21. American Pain Society. Pediatric chronic pain: A position statement from the American Pain Society. [Accessed July 25, 2005]. http://www.ampainsoc.org/advocacy/pediatric.htm
22. Mehling WE, Gopisetty V, Daubenmier J, Price CJ, Hecht FM, Stewart A. Body awareness: construct and self-report measures. PLoS One 2009;4(5):e5614. [PubMed: 19440300]
23. Cremeens J, Eiser C, Blades M. Brief report: assessing the impact of rating scale type, types of items, and age on the measurement of school-age children’s self-reported quality of life. J Pediatr Psychol 2007;32(2):132–138. [PubMed: 16625022]
24. Cleeland CS, Ryan KM. Pain assessment: global use of the Brief Pain Inventory. Ann Acad Med 1994;23(2):129–138.
25. Daut RL, Cleeland CS, Flanery RC. Development of the Wisconsin Brief Pain Questionnaire to assess pain in cancer and other diseases. Pain 1983;17(2):197–210. [PubMed: 6646795]
26. Jensen MP, Turner JA, Romano JM, Fisher LD. Comparative reliability and validity of chronic pain intensity measures. Pain 1999;83(2):157–162. [PubMed: 10534586]
27. Jensen, MP.; Karoly, P. Self-report scales and procedures for assessing pain in adults. In: Turk, DC.; Melzack, R., editors. Handbook of Pain Assessment. New York: Guilford; 1992. p. 132-137.
28. Rosenbaum PL, Palisano RJ, Bartlett DJ, Galuppi BE, Russell DJ. Development of the Gross Motor Function Classification System for cerebral palsy. Dev Med Child Neurol 2008;50(4):249–253. [PubMed: 18318732]
29. Morris C, Bartlett D. Gross Motor Function Classification Systems: impact and utility. Dev Med Child Neurol 2004;46(1):60–65. [PubMed: 14974650]
30. Chambers CT, Reid GJ, Craig KD, McGrath PJ, Finley GA. Agreement between child and parent reports of pain. Clin J Pain 1998;14(4):336–342. [PubMed: 9874013]
31. Palermo TM, Zebracki K, Cox S, Newman AJ, Singer NG. Juvenile idiopathic arthritis: parent-child discrepancy on reports of pain and disability. J Rheumatol 2004;31(9):1840–1846. [PubMed: 15338510]
32. Ehde D, Jensen M, Engel J, Turner J, Hoffman A, Cardenas D. Chronic pain secondary to disability: a review. Clin J Pain 2003;19(1):3–17. [PubMed: 12514452]
33. Molton IR, Stoelb BL, Jensen MP, Ehde DM, Raichle KA, Cardenas DD. Psychosocial factors and adjustment to chronic pain in spinal cord injury: replication and cross-validation. J Rehabil Res Dev 2009;46(1):31–42. [PubMed: 19533518]
34. Ullrich PM, Jensen MP, Loeser JD, Cardenas DD, Weaver FM. Pain among veterans with spinal cord injury. J Rehabil Res Dev 2008;45(6):793–800. [PubMed: 19009466]
35. Molton I, Jensen MP, Ehde DM, Carter GT, Kraft GH, Cardenas D. Coping with chronic pain among younger, middle-aged, and older adults living with neurologic injury and disease: a role for experiential wisdom. J Aging Health 2008;20(8):972–996. [PubMed: 18791184]
36. Ehde DM, Osborne TL, Hanley MA, Jensen MP, Kraft GH. The scope and nature of pain in persons with multiple sclerosis. Mult Scler 2006;12(5):629–638. [PubMed: 17086910]
37. Jensen MP, Hoffman AJ, Cardenas DD. Chronic pain in individuals with spinal cord injury: a survey and longitudinal study. Spinal Cord 2005;43(12):704–712. [PubMed: 15968299]

38. Huguet A, Miró J. The severity of chronic pain in children and adolescents: an epidemiological study. J Pain 2008;9(3):226–236. [PubMed: 18088558]
39. Palermo TM. Impact of recurrent and chronic pain on child and family daily functioning: a critical review of the literature. J Dev Behav Pediatr 2000;21(1):58–69. [PubMed: 10706352]
40. Hunfeld JA, Perquin CW, Duivenvoorden HJ, et al. Chronic pain and its impact on quality of life in adolescents and their families. J Pediatr Psychol 2001;26(3):145–153. [PubMed: 11259516]
41. Strax TE. Psychological issues faced by adolescents and young adults with disabilities. Pediatr Ann 1991;20(9):507–511. [PubMed: 1834979]
42. Rudich Z, Lerman SF, Gurevich B, Weksler N, Shahar G. Patients' self-criticism is a stronger predictor of physician's evaluation of prognosis than pain diagnosis or severity in chronic pain patients. J Pain 2008;9(3):210–216. [PubMed: 18055267]
43. Beach MC, Roter DL, Wang NY, Duggan PS, Cooper LA. Are physicians' attitudes of respect accurately perceived by patients and associated with more positive communication behaviors? Patient Educ Couns 2006;62(3):347–354. [PubMed: 16859867]
44. Oberlander TF, Gilbert CA, Chambers CT, O'Donnell ME, Craig KD. Biobehavioral responses to acute pain in adolescents with a significant neurologic impairment. Clin J Pain 1999;15(3):201–209. [PubMed: 10524473]
45. Hinton VJ, Nereo NE, Fee RJ, Cyrulnik SE. Social behavior problems in boys with Duchenne muscular dystrophy. J Dev Behav Pediatr 2006;27(6):470–476. [PubMed: 17164619]
46. Lester N, Lefebvre JC, Keefe FJ. Pain in young adults—III: relationships of three pain-coping measures to pain and activity interference. Clin J Pain 1996;12(4):291–300. [PubMed: 8969874]
47. Palermo TM, Witherspoon D, Valenzuela D, Drotar D. Development and validation of the Child Activity Limitations Interview: a measure of pain-related functional impairment in school-age children and adolescents. Pain 2004;109(3):461–470. [PubMed: 15157707]
48. Eddy LL, Engel JM. The impact of child disability type on the family. Rehabil Nurs 2008;33(3): 98–103. [PubMed: 18517144]
49. McDonald CM, Abresch RT, Carter GT, et al. Profiles of neuromuscular disease. Duchenne muscular dystrophy. Am J Phys Med Rehabil 1995;74(5 suppl):S70–S92. [PubMed: 7576424]
50. McDonald CM, Abresch RT, Carter GT, Fowler WM Jr, Johnson ER, Kilmer DD. Profiles of neuromuscular disease. Becker muscular dystrophy. Am J Phys Med Rehabil 1995;74(5 suppl):S93–S103. [PubMed: 7576425]
51. Johnson ER, Abresch RT, Carter GT, et al. Profiles of neuromuscular disease. Myotonic muscular dystrophy. Am J Phys Med Rehabil 1995;74(5 suppl):S104–S116. [PubMed: 7576418]
52. McDonald CM, Johnson ER, Abresch RT, Carter GT, Fowler WM Jr, Kilmer DD. Profiles of neuromuscular disease. Limb-girdle syndromes. Am J Phys Med Rehabil 1995;74(5 suppl):S117–S130. [PubMed: 7576419]
53. Kilmer DD, Abresch RT, McCrory MA, et al. Profiles of neuromuscular disease. Facioscapulohumeral dystrophy. Am J Phys Med Rehabil 1995;74(5 suppl):S131–S139. [PubMed: 7576420]
54. Carter GT, Abresch RT, Fowler WM Jr, Johnson ER, Kilmer DD, McDonald CM. Profiles of neuromuscular disease. Hereditary motor and sensory neuropathy, types I and II. Am J Phys Med Rehabil 1995;74(5 suppl):S140–S149. [PubMed: 7576421]
55. Carter GT, Abresch RT, Fowler WM Jr, Johnson ER, Kilmer DD, McDonald CM. Profiles of neuromuscular disease. Spinal muscular atrophy. Am J Phys Med Rehabil 1995;74(5 suppl):S150–S159. [PubMed: 7576422]
56. Carter GT. Phrenic nerve involvement in Charcot-Marie-Tooth. Muscle Nerve 1995;18(10):1215–1216. [PubMed: 7659121]
57. Carter GT, Bird TD. Ventilatory support in facioscapulohumeral muscular dystrophy. Neurology 2005;64(2):401–402. [PubMed: 15668464]
58. Carter GT, Kilmer DD, Bonekat HW, Lieberman JS, Fowler WM Jr. Evaluation of phrenic nerve and pulmonary function in hereditary motor and sensory neuropathy, type I. Muscle Nerve 1992;15(4):459–462. [PubMed: 1565114]
59. Smith MT, Perlis ML, Smith MS, Giles DE, Carmody TP. Sleep quality and presleep arousal in chronic pain. J Behav Med 2000;23(1):1–13. [PubMed: 10749008]

60. Gold JI, Mahrer NE, Yee J, Palermo TM. Pain, fatigue, and health-related quality of life in children and adolescents with chronic pain. Clin J Pain 2009;25(5):407–412. [PubMed: 19454874]

Table 1

Pain Frequency (n = 42)

	Frequency	Percentage
None of the time	10	24
Once or twice/week	12	28
A few times/week	11	26
Fairly often	4	10
Very often	2	5
Everyday or almost everyday	3	7

Table 2

Pain Location and Numerical Rating Scale (NRS) Pain Intensity (n = 23)

Location	N (%)	Min	Max	Mean	SD
Legs	23 (100)	1	8	5.36	1.86
Back	15 (65)	1	7	3.87	1.77
Head	13 (57)	1	6	3.69	1.65
Bottom/hips	13 (57)	1	5	3.31	1.55
Feet	13 (57)	1	10	4.85	2.76
Neck	10 (44)	2	5	3.40	1.17
Shoulders	10 (44)	2	7	3.50	1.65
Arms	6 (26)	1	4	2.33	1.03
Belly/pelvis	6 (26)	1	9	4.17	3.06
Chest	7 (30)	1	10	3.43	3.04
Hands	4 (17)	2	6	3.00	2.00

Table 3

Pain Interference With Life Domains (n = 23)

	N (%)	Min	Max	Mean	SD
Sleep	14 (61)	1	8	3.36	2.17
General activity	14 (61)	1	6	3.29	1.54
Mood	13 (57)	1	7	2.92	1.60
Mobility (ability to get around)	12 (52)	1	6	3.83	1.74
School/work	12 (52)	1	10	4.08	2.93
Recreational activities	11 (48)	1	7	3.45	1.97
Self-care (taking care of daily needs)	9 (39)	1	7	2.89	1.83
Learning new information or skills	8 (35)	1	9	3.50	2.88
Relations with other people	8 (35)	1	6	3.13	1.46
Communication with others	8 (35)	1	5	2.50	1.41
Enjoyment of life	7 (30)	1	5	3.57	1.62
Social activities	7 (30)	1	7	3.57	2.44
Communication with others	8 (35)	1	5	2.50	1.41

Table 4

Factors Exacerbating Pain

Factor	N (%)
Exercise	11 (48%)
Walking	7 (30%)
Stress	6 (26%)
Stretching	8 (35%)
Wearing a splint/prosthesis	0 (0%)

Table 5

Factors Relieving Pain

Factor	N (%)
Relaxing activities	14 (61%)
Rest/sleep	12 (52%)
Medication	11 (48%)
Stretching	10 (44%)
Exercise	7 (30%)

Table 6

Frequency and Helpfulness of Pain Treatment

Intervention	Ever Used, N (%)	Currently Using, N (%)	Helpfulness[a] Mode (range)
Nonsteriodal anti-inflammatories	19 (83)	11(57)	3.0 (0–5)
Acetaminophen	19 (83)	11 (57)	3.0 (0–5)
Physical or occupational therapy	12 (52)	7 (58)	3.0 (0–5)
Tricyclic antidepressants	4 (17)	2 (50)	2.0 (0–5)
Opioid medications	4 (17)	2 (50)	4.5 (3–5)
Counseling/psychotherapy	2 (9)	1 (50)	4.5 (2–5)
Therapeutic massage	1 (4)	1 (50)	5
TENS	0 (0)	0 (0)	0

Abbreviation: TENS, transcutaneous electrical nerve stimulation.

[a] 0 = not at all, 5 = extremely helpful.